



SEVERITY OF JOINT INVOLVEMENT USING HEMOPHILIA JOINT HEALTH SCORE 2.1 (HJHS) IN CHILDREN SUFFERING FROM HEMOPHILIA A

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Abstract:

Introduction: Hemophilia A is a rare, recessive X-linked, congenital disorder caused by deficient or defective coagulation factor VIII (FVIII) and the deficiency of this factor leads to excessive bleeding following trauma or injury and spontaneous bleeding episodes, primarily in the joints, muscles, and soft tissues. Worldwide the diagnosis and treatment provisions vary widely. In developing countries, access to health care is very limited and very few people can be tested for hemophilia. Developed countries, on the other hand, use personalized prophylactic therapy to optimize outcomes and work towards zero bleeds. Numerous measuring methods for the evaluation of hemophilic have been developed. One of the most used systems is the Hemophilia Joint Health Score (HJHS) 2.1 given its sensitivity to clinical changes appearing in the joints because of recurrent hemarthrosis.

Material and Methods: Severity of joint involvement using Hemophilia joint Health score 2.1 was measured by various parameters. Nine items are assessed to evaluate the status of a joint which include swelling, duration of swelling, muscle atrophy, crepitus of motion, range of motion (extension and flexion loss), joint pain, strength, and gait. The summation of the 9 HJHS items in 6 index joints provides users with a relative indicator of joint health. Lower HJHS score represents superior joint health.

Results: A total of 51 children were included in the study. Mean HJHS was highest in 10-14 years group while it was lowest (2.81) in 6-9 years group. It was shown that score increases with age indicating that disability increases with the age. Swelling and flexion loss was contributing highest among the all disability items taken into account for calculation of HJHS. It was also observed that almost half of the score was due to contribution of swelling and flexion loss. It was observed that man HJHS was higher in cases diagnosed after the age of 12 months (2.68) as compared to cases that were diagnosed earlier (7.96). This difference was statistically significant ($P=0.049$).

Conclusion: Comprehensive musculoskeletal assessment for monitoring joint health in hemophilia can be done by HJHS. It showed moderate-to-strong correlation across the age group and cases diagnosed after the age of 12 months.

keywords: Hemophilia, HJHS 2.1, musculoskeletal assessment.

Introduction

Hemophilia A is a rare, recessive X-linked, congenital disorder caused by deficient or defective coagulation factor VIII (FVIII) and the deficiency of this factor leads to excessive bleeding following trauma or injury and spontaneous bleeding episodes, primarily in the joints, muscles, and soft tissues^{i, ii}. The incidence of hemophilia is very low and the World Foundation of Hemophilia which was founded in 1963, has used a multidisciplinary worldwide approach to improve care for patients with the disease. In children with severe hemophilia, in addition to the management of bleeding episodes, primary prophylaxis, is the recommended treatment strategyⁱⁱⁱ. In hemophilia

management development of inhibitors, the alloantibodies to factor concentrates, can cause severe complications. Once a patient develops inhibitors, he will experience more frequent and severe bleeding episodes and will need more factor concentrates or bypassing agents^{iv}.

Worldwide the diagnosis and treatment provisions vary widely. In developing countries, access to health care is very limited and very few people can be tested for hemophilia. Developed countries, on the other hand, use personalized prophylactic therapy to optimize outcomes and work towards zero bleeds^v.

In comprehensive care of hemophilia the treatment and prevention of bleeding is important, the long-

term management of hemophilic arthropathy and other bleeding complications are recognised, the management of significant complications from treatment is done^{vi}.

Numerous measuring methods for the evaluation of hemophilic have been developed. One of the most used systems is the Hemophilia Joint Health Score (HJHS) 2.1 given its sensitivity to clinical changes appearing in the joints because of recurrent hemarthrosis^{vii}.

MATERIAL AND METHODS

Present study was carried out in the New Civil Hospital, Surat Gujarat. This prospective study was conducted on the patients suffering from Hemophilia A from Surat District.

Severity of joint involvement using Hemophilia joint Health score 2.1 was measured by various parameters.

The Hemophilia Joint Health Score (HJHS) was developed to assess joint health in people with hemophilia. HJHS shows a measure of joint health. Nine items are assessed to evaluate the status of a joint which include swelling, duration of swelling, muscle atrophy, crepitus of motion, range of motion (extension and flexion loss), joint pain, strength, and gait. The summation of the 9 HJHS items in 6 index joints provides users with a relative indicator of joint health. Lower HJHS score represents superior joint health. HJHS scores may assist in the development of individualized musculoskeletal treatment plans or programs. The HJHS is an internationally validated physical examination assessment tool with excellent interobserver and test-retest reliability^{viii}.

RESULTS

A total of 51 children's were included in the study.

Table 1: Age wise distribution and correlation of HJHS with age of the cases

Age in years	Cases	Percentage	HJHS Mean	SD	P value (t test)
4-5	6	11.8%	1.17	2.86	0.131
6-9	21	41.2%	2.81	6.02	
10-14	24	47.1%	8.00	12.87	
Total	51	100%	5.06	9.98	

Mean HJHS was highest in 10-14 years group while it was lowest (2.81) in 6-9 years group. It was shown that score increases with age indicating that disability increases with the age.

Table 2: Individual Joint and Item wise score

Item	Joint (Sum of score of all cases)						Total	Percentage
	LE	RE	LK	RK	LA	RA		
Swelling	0	9	14	18	8	8	57	24.1
Duration	0	0	4	5	3	2	14	5.9
Muscle atrophy	0	0	2	4	1	1	8	3.4
Crepitus on motion	1	7	10	12	4	3	37	15.6
Flexion loss	1	13	12	15	7	8	56	23.6
Extension loss	1	9	8	10	3	4	35	14.8
Joint pain	2	4	5	7	4	4	26	11.0
Strength	0	0	1		1	1	4	1.7
Total	5	42	56	1	31	31	237	100

*LE = Left Elbow, RE=Right Elbow, LK=Left Knee,RK=Right Knee , LA=Left Ankle,RA=Right Ankle.

In above table it was depicted that based on the total of item wise score swelling and flexion loss was contributing highest among the all disability items taken into account for calculation of HJHS. It was also observed that almost half of the score was due to contribution of swelling and flexion loss.

Table 3: Correlation of HJHS with habit of exercise

Exercise Habit	Frequency	HJHS Mean	SD	
Yes	27	6.41	10.69	0.311
No	24	3.54	9.10	
Total	51	5.06	9.98	

Mean HJHS was higher in those having habit of exercise (6.41) compared to cases who did not do exercise (3.54). However application of statistical test indicated that this difference was not statically not significant ($p>0.05$),so HJHS was not correlated with the habit of exercise.

Table4: Correlation of HJHS with age of diagnosis

Age of diagnosis	Frequency	HJHS mean	SD	P value (t test)
< 12 months	23	7.96	10.37	0.049
≥ 12 months	28	2.68	9.15	
Total	51	5.06	9.98	

It was observed that man HJHS was higher in cases diagnosed after the age of 12 months (2.68) as compared to cases that were diagnosed earlier (7.96). This difference was statistically significant ($P=0.049$). So HJHS and thus disability was higher in cases whom the disease was diagnosed earlier.

Table: 5 Correlation of HJSS with treatment modality

Treatment modalities	Frequency	HJHS	SD	P value (t test)
Factor VIII	15	2.53	7.55	0.247
Factor VIII and other	36	6.11	10.75	
Total	51	5.06	9.98	

As shown in the table mean HJHS was higher in cases receiving factor VIII with other drugs(6.11)compare to cases receiving factor VIII only (2.53). However application of statistically not significant ($p>0.05$) ,so HJHS was not correlated with the treatment morality.

Discussion:

In this study it was observed that mean HJHS was highest in 10-14 years group while it was lowest (2.81) in 6-9 years group. It was shown that score increases with age indicating that disability increases with the age. In a study by McLaughlin P^{ix} et al in 2018 it was shown that Data from 120 patients with a median age 33 years (range 19-73) were included. Median total HJHS was 19, increasing with age: 18-30 years-7, 31-50 years-25 and 51-73 years-44. Similarly, median total HAL score was 80 with decreased function associated with increasing age: 18-30 years-90.4, 31-59 years-71.7, 51-73 years-49.5. Median Total HJHS and HAL demonstrated strong correlation ($r_s = 0.66$, $P < 0.01$). Moderate-to-strong correlations were seen across the entire age group between the HJHS.

It was observed that mean HJHS was higher in cases diagnosed after the age of 12 months (2.68) as compared to cases that were diagnosed earlier (7.96). This difference was statistically significant ($P=0.049$). So HJHS and thus disability was higher in cases whom the disease was diagnosed earlier. Mean HJHS was in this study was higher in cases residing more than 15 km away from the hospital (8.23 ± 13.386) as compared to cases nearer to the hospital (3.97 ± 8.468). However the difference was not statistically significant. So HJHS was not correlated with the distance from home to hospital.

In a study it differences in preference weights for HJHS items highlight the potential under- or overestimation of true joint health using the current ordinal scoring system. Also they concluded that an updated scoring system using weighted items may improve the precision of HJHS assessment, leading to improved clinical management of joint health, while providing a robust research tool^x.

Conclusion

Comprehensive musculoskeletal assessment for monitoring joint health in haemophilia can be done by HJHS. It showed moderate-to-strong correlation across the age group and cases diagnosed after the age of 12 months. More prospective studies are required in the area to better understand the clinical utility of HJHS.

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